TESTIMONY OF

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Concerning
Chronic Wasting Disease

Before the
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COMMITTEE ON RESOURCES
UNITED STATES HOUSE OF REPRESENTATIVES

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Good morning, Mr. Chairman and committee members. I am Michael Miller, Staff Veterinarian for the Colorado Division of Wildlife. I sincerely appreciate the invitation to appear before your Subcommittees to provide some background information on chronic wasting disease of deer and elk as a foundation for today's discussions. I'm sure that all of you, as members of the Resources Committee, are at least somewhat familiar with this disease, so much of this may well be review. In the next few minutes I plan to briefly review key features of chronic wasting disease, its history, and what we currently know about its distribution and occurrence in both free-ranging and farmed deer and elk.

The information I'm presenting is a synthesis of data, much of it previously published in peer-reviewed scientific journals. These data have been generated by a number of talented state, federal, and university scientists who have been collaboratively working on chronic wasting disease for over two decades. In particular, please recognize the contributions that Dr. Beth Williams from the University of Wyoming, Dr. Terry Spraker from Colorado State University, Dr. Katherine O'Rourke and colleagues from the USDA ARS laboratory in Pullman, Washington, Dr. Byron Caughey and colleagues from the NIH Rocky Mountain Laboratory in Hamilton, Montana, and Dr. Tom Thorne from the Wyoming Game and Fish Department have made to our collective understanding of chronic wasting disease in deer and elk.

The prion diseases

Chronic wasting disease, or CWD, is one of a relatively small but important group of diseases called transmissible spongiform encephalopathies (or TSEs, or prion diseases). These diseases are believed to be caused by a number of unique strains of infectious, self-propagating prion protein. The prion diseases are relatively new to science, and many aspects of their biology are poorly understood. Some of the known prion diseases affect domestic or wild animals, while others occur naturally in humans.

There are three important prion diseases of food-producing animals. The most common and widespread of these is scrapie of domestic sheep and goats, which occurs throughout the United States and virtually world-wide and has been recognized as an animal health problem for over 200 years. Chronic wasting disease is relatively rare, affecting native North American deer and elk species in the cervid (or deer) family -- we'll spend most of our time today talking more about CWD. However, the third of these is the one that focused public attention on the prion diseases: I'm sure you've all heard of bovine spongiform encephalopathy, also called BSE or "mad cow disease", and are well aware of the impacts that this disease has had on agricultural economies in the United Kingdom and several European countries. I'm sure you're also aware that, in a relatively small but still significant number of cases, BSE apparently was transmitted to humans and manifested itself as a variant of Creutzfeldt-Jakob disease (or vCJD). Although sporadic CJD occurs in human populations world-wide with an attack rate of 1-2 cases/million people, the occurrence of variant CJD in British citizens sparked a public health crisis, perceived if not real, that has influenced public perceptions about animal prion diseases both overseas and here in the US.

It's very important for all of you to understand that chronic wasting disease is ***NOT*** simply BSE in deer and elk, as some might have you believe. Although these maladies are in the same disease family, we know that the strain of prion that appears to cause CWD is quite different from the strain that causes BSE, and also appears somewhat different from strains of scrapie that naturally infect domestic sheep and goats here in the US and overseas. The true origin of CWD remains unknown (despite what you may read on the Web or in the newspaper) -- whether it began as scrapie or as a sporadic disease of deer or elk is, and probably will always be, a mystery.

Chronic wasting disease

The known natural host range for CWD is limited to three species, all from the"deer" (or cervid) family: mule deer and white-tailed deer (both in the genus *Odocoileus*), and elk (in the genus *Cervus*). All three natural host species show similar susceptibility to CWD, but there appear to be some species- or genus-specific differences that may influence how the disease behaves in each species. Chronic wasting disease does not appear to be naturally transmissible to domestic livestock or pets, other wildlife species, or humans. Experimentally, chronic wasting disease has been transmitted to several species using unnatural

exposure routes. In general, experimental transmission of the CWD prion is much less efficient than experimental transmission of scrapie. After over two centuries of experience with scrapie, this more common animal prion disease has not been shown to transmit naturally to species other than sheep and goats; this offers considerable reassurance that the natural host range of CWD is likely to be limited only to select species in the deer family.

The hallmark clinical signs for end-stage CWD are emaciation and abnormal behavior. In practice, subtle changes in behavior, attentiveness, and locomotion can be seen in most infected animals some months before end-stage disease develops. Other health problems, particularly pneumonia and injury, can confound the diagnosis of CWD in deer and elk. Consequently, as with other animal TSEs, laboratory diagnosis is necessary to confirm infections in suspect animals.

There are several important epidemiological features of CWD. We know to expect a prolonged incubation period in exposed deer and elk that averages somewhere in the range of 20-30 months with natural infections, but may be somewhat shorter or considerably longer (perhaps 60 months or more) in individual cases. Susceptibility to CWD infection appears to be relatively uniform among species, between sexes, and across age classes; there does appear to be some genetic influence on susceptibility in elk but not deer. CWD appears to be maintained naturally in both captive and free-ranging cervid populations; epidemics persist in the absence of exposure to contaminated feeds or other likely outside sources of infection. Direct or indirect animal-to-animal transmission, not necessarily along maternal lines, drives CWD epidemics. Although we don't know precisely how CWD is transmitted among deer and elk, the agent is probably shed in feces, saliva, and perhaps urine. In addition, contaminated environments likely play a role in epidemics and the recurrence of disease in some situations -- in some cases, the CWD agent apparently persisted in heavily contaminated environments for years after all infected deer or elk had been removed. This environmental persistence obviously represents a significant obstacle for eradicating CWD in places where it is already well-established, either in captivity or in the wild.

Chronic wasting disease is not a new prion disease. The clinical syndrome of "chronic wasting" was first recognized in captive mule deer in Colorado in the late 1960s, but tying the first recognition of a disease like this to its first occurrence seems like a substantial leap of faith. Based on what we now know about its distribution and occurrence, it is quite plausible that CWD actually arose in captive and/or free-ranging cervids 40 or more years ago. In 1977, Drs. Williams and Young recognized that CWD was, in fact, a TSE. Within a few years of finally having a clear-cut diagnostic criterion, CWD was first detected in free-ranging elk and deer in northestern Colorado and southeastern Wyoming – however, these were almost certainly not the first cases to occur in the wild in either state. Similarly, the first diagnosis of CWD in a farmed elk was made in Saskatchewan in 1996 – in retrospect, this most assuredly was not the first case to occur in the elk industry in either Canada or the US because Canadian investigations have shown that some infected elk were apparently imported into Canada from South Dakota in the late 1980s, if not earlier. In the last 2 years, CWD has now been detected in free-ranging deer in several locations well outside the original known endemic focus of southeastern Wyoming and northeastern Colorado. Precisely how and when these scattered foci of infection came about is not entirely clear. So, although there may be some value in recognizing these milestones in the history of CWD, it's important not to interpret these as absolute timelines for the emergence of this disease.

Approaches for assessing status

Before proceeding into what we know about the status of CWD in both free-ranging and farmed cervids, I

want to provide a bit of context on how we've come to know what we know. To truly appreciate how much we know about CWD, it's important to compare approaches for detecting CWD with traditional approaches for detecting TSEs in other animal species. For both scrapie and BSE, there has tended to be a focus on clinical cases as the means of detecting new infections. This is clearly an effective approach, particularly when such diseases are reportable, but there are biases and limitations inherent in this strategy. Moreover, these surveillance programs are often conducted in an atmosphere where there may be substantial economic penalties for owners of infected animals or herds.

Please recognize that animals showing end-stage clinical disease represent the "tip of the iceberg" with respect to the overall rate of infection in the population of interest. Those of us working with CWD also recognized this some time ago, and modified our surveillance strategies accordingly. Initially, we used histopathology of brainstem samples as an adjunct to surveying populations for CWD, and gained a much better appreciation of the size and shape of the iceberg. Beginning in 1996, we adopted immunohistochemistry (IHC) of brainstem as our screening tool for surveillance, and again improved our appreciation of the iceberg's depth and magnitude. And, for the last 3 years, we've been able to use IHC of tonsillar tissues to gain an almost , but not quite complete, picture of the CWD iceberg. We know even these IHC-based estimates of CWD prevalence are still a little low, but they're much closer to truth than data generated otherwise.

Surveillance systems for CWD in free-ranging wildlife evolved in the absence of regulatory or economic pressure. To date, the motivations for reliably estimating the distribution and prevalence of CWD in native wildlife populations have been twofold: scientific curiosity, accompanied by a sense of responsibility for acquiring *and conveying to the public* accurate information about this disease and its occurrence in public resources. Similarly, the farmed elk industry recognized early on the value of detecting CWD in their herds as a basis for effective disease management. In this environment, three somewhat distinct approaches to CWD surveillance have evolved and are currently in use in varying combinations. An appreciation of the details and applications of each is important in interpreting data on CWD status.

The foregoing caveats notwithstanding, surveillance for clinical suspects remains an effective tool for detecting new foci of CWD infection in both captive and free-ranging settings. Under these systems, clinical suspects are sampled whenever available. Histopathology of brainstem is usually sufficient to diagnose cases, but IHC is a valuable adjunct in many cases. Data are clearly biased, and consequently are of little use in estimating prevalence. This approach is very similar to traditional scrapie surveillance in the US.

In some captive settings, CWD surveillance has been applied to all natural mortalities, and in some cases to all mortalities regardless of proximate cause. Several states have adopted this approach in rules that regulate their elk industries. Mortality-based surveillance is also an effective tool for detecting new foci of CWD infection, and has resulted in the disclosures of several infected elk farms over the last 3 years. Here again, histopathology of brainstem (a specific portion called the "obex") is usually sufficient to diagnose cases, but IHC is a valuable adjunct. Inherent biases in these data limit their use in estimating prevalence. This approach is considerably more aggressive than traditional scrapie surveillance in the US, and has facilitated CWD detection in the elk industry.

And finally, those of us investigating CWD in free-ranging populations over the last decade have developed efficient techniques for conducting geographically-targeted random sampling of harvested deer and elk to

estimate prevalence and monitor trends. In these surveys, sections of obex and, more recently, tonsil are collected and examined via immunohistochemistry; infections can be staged further by histopathology. Data from these samples represent relatively unbiased point estimates of CWD prevalence. Unfortunately, comparable slaughter survey data for scrapie and BSE have not been reported formally, confounding comparisons of epidemic severity between CWD and, for example, scrapie in the US. This lack of comparable data has perhaps fostered misperceptions about CWD.

Current status in free-ranging and captive deer and elk

Using various combinations of these 3 surveillance approaches, we have developed a good basic understanding of CWD's status in North America. At present, there appear to be 2 relatively distinct CWD epidemics occurring in North American cervid populations. One epidemic focus is in free-ranging cervids in southeastern Wyoming, northeastern Colorado, and the southwestern corner of the Nebraska panhandle. The other epidemic is occurring in a relatively small number of farmed elk herds scattered across the US and Canada, with apparent spill-over to local populations of free-ranging deer in Saskatchewan, northwestern Nebraska, and South Dakota. Although a common source for both epidemics has been speculated, and would certainly be the most parsimonious way to explain the origins of CWD, no common thread linking all of these has been demonstrated to date.

Alternatively, not knowing how or when CWD originated in the first place, it is conceivable that whatever event gave rise to CWD once could have occurred again in farmed elk, and that the two epidemics are not directly related. This also could perhaps explain the recent occurrences of CWD in Wisconsin and northwestern Colorado, where no known exposure source has been identified to date. Regardless of whether or not they have a common root origin, these epidemics, as we understand them today, appear to be essentially unrelated epidemiologically and are probably best considered independently.

I'll first give some highlights of the main CWD epidemic in free-ranging deer and elk.

Chronic wasting disease has been recognized in free-ranging deer and elk since the early 1980s. This epidemic is likely related in some way to cases originally reported in captive research animals, but which came first is truly a chicken-or-egg question that we're probably never going to answer. Initially, clinical cases were recognized in both free-ranging deer and elk in northeastern Colorado and southeastern Wyoming. Surveillance for clinical suspects has been ongoing in both states since these first cases were detected. Harvest-based surveys were conducted intermittently beginning in 1983, and annually in some areas beginning in 1990. Since 1996, we've been using immunohistochemistry to enhance detection of infected animals. In all, about 15,000 deer and elk harvested or culled in northeastern Colorado, southeastern Wyoming, and western Nebraska have been sampled to date. Ongoing random surveys have revealed a contiguous endemic focus of CWD that primarily involves mule deer, but also includes white-tailed deer and elk where they occur in that area. This so-called "endemic area" spans over 16,000 mi^2 of mixed native habitats. The most intensively infected area extends from the Laramie Mountains in Wyoming south into the northern Front Range in Colorado – in this area, average infection rates exceed 10% in sampled mule deer. This high prevalence ridge is surrounded by areas of successively lower prevalence. On the fringes of the endemic area, prevalence is 1% or less in deer. Elk reside in the western half of this area; CWD prevalence in this species is <1% in all of the areas sampled. Where white-tailed deer ranges overlap with mule deer, CWD prevalence is similar in both deer species. The spatial pattern of relative disease prevalence strongly suggests that what we're seeing is actually an epidemic occurring in slow motion, extending geographically through natural animal movements. Computer models suggest that CWD has

likely been present in some of the more heavily infected areas for 35 years or more. Surveys have been conducted over the last 5 years in other parts of Colorado and Wyoming, as well as in portions of a number of nearby and distant states and provinces by responsible wildlife management and animal health agencies. With a few notable exceptions that you'll hear more about later, none of these surveys have revealed other foci of CWD presently exist in free-ranging cervids. In all, over 10,000 deer and elk from these areas had tested negative through the 2000-2001 sampling season. At the very least, these data clearly demonstrate that CWD is not uniformly prevalent across all of our native cervid populations in the US and Canada.

I now want to turn your attention briefly to the status of CWD in farmed elk.

As is the case with CWD in free-ranging cervids, this disease has likely been in the captive wildlife industry for some time. The first report of a farmed elk with CWD came from Saskatchewan in 1996 – it was a routine submission of an animal with chronic pneumonia that was refractory to treatment. Nearly 2 years later, a similar case was submitted from a South Dakota elk farm. Subsequent epidemiological investigations, as well as submissions made voluntarily or in compliance with mandatory surveillance rules, have led to disclosure of CWD infections in 62 game farms in 6 western states and 2 Canadian provinces; a few others remain under investigation. South Korea also apparently received infected elk from Saskatchewan in 1997, representing the first known extension of CWD beyond the North American continent. To date, all of the CWD cases in privately-owned cervids have occurred in elk. However, very little surveillance has been conducted in privately-owned captive deer herds to date. The apparent intensity of infections in herds studied to date has varied widely, and probably reflects influences of husbandry, as well as the duration of infection in a particular herd. There are epidemiological connections, documented through animal movements, among some but not all of the infected herds. And, although there is geographic overlap between the location of some infected Colorado farms and the endemic focus in free-ranging deer, with one exception epidemiological investigations really don't support free-ranging deer as the most likely source of infection in these cases.

The elk industry, in conjunction with responsible state and federal animal health agencies, has been quite aggressive in dealing with CWD. Of the infected elk farms identified to date, only a few remain under some form of quarantine and negotiated disease management – the remaining herds either have been or are in the process of being depopulated. Before federal funds were available through a national CWD program for captive elk in the US, many of the herds were depopulated voluntarily or with the help of industry funding. Both Canada and the US are in the process of developing national CWD programs. In the interim, several states have programs and regulations for CWD in farmed cervids in various stages of development and implementation.

Managing chronic wasting disease

Although CWD is of understandable concern to a variety of interests, there is a considerable amount of information available to help assess risk and guide policy decisions. In the US, CWD is probably best viewed as 2 separate epidemics, one involving free-ranging cervids and the other involving captive elk. Neither of these epidemics is particularly new. Both epidemics are relatively well-described, particularly in comparison to scrapie in the US. CWD is naturally maintained in both free-ranging and captive cervid populations, and thus management will be challenging in both settings. In the short-term, CWD in captive elk is much more likely to be manageable than CWD in free-ranging cervids. I have no doubt, however, that new knowledge on CWD and other TSEs will factor into future plans for further understanding and managing both problems.

There are perceptions that CWD may somehow threaten human or traditional domestic livestock health. These perceptions clearly factor into motivations for managing CWD, even though data and experiences to date suggest those threats appear vanishingly small. We know, however, that CWD represents a significant threat to the health and long-term stability of free-ranging deer or elk resources that are an important component of both the ecology and economy of virtually every state represented in this Congress. According to published model forecasts of CWD epidemics in deer populations, unmanaged outbreaks will likely devastate infected herds over a period of several decades. In the interim, the public value of these infected herds may well be diminished, simply by virtue of their status as CWD-infected. At present, the threat posed to our valuable wildlife resources seems more than sufficient to justify more aggressive actions, fostered by state-federal cooperation, to identify foci of infection, contain these and, where possible, eliminate chronic wasting disease from our nation's free-ranging deer and elk populations.

Respectfully submitted,

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